Prolonged QT Syndromes

Prolonged (long) QT syndromes (LQTS) can be congenital or acquired. Syncope is the hallmark of the inherited forms of prolonged QT syndrome. These events are commonly associated with stress, emotion, exercise, or other situations associated with sympathetic stimulation. A rare autosomal recessive form of prolonged QT syndrome, called Jervell Lange-Nielsen syndrome, is associated with congenital deafness. Acquired iatrogenic prolonged QT syndrome is far more common than inherited forms of LQTS.

ANESTHETIC CONSIDERATIONS:

1. Potential for hemodynamic collapse secondary to rapid polymorphic VT and Vfib
2. Avoid further prolongation of the QTc
    - Sympathetic stimulation
    - Medications
    - R stellate ganglion block
    - L stellate ganglion stimulation
    - Electrolyte disturbances (hypoK, hypoMg, hypoCa)
3. Risk of sudden cardiac death → May have pacemaker / AICD
4. Potential for pre-operative optimization with β-blockade or left stellate ganglion block

ANESTHETIC GOALS:

- Avoid triggers of prolonged QT (emotional stress, hypokalemia, bradycardia, sympathetic nervous system stimulation, drugs)
- Anticipate and prepare for potential hemodynamic consequences of rapid polymorphic VT

DEFINITIONS

- a prolongation of the QTc exceeding 460 (men) to 480 (women) ms

HISTORY & PHYSICAL

- Congenital long QT syndrome is mostly an autosomal dominant disorder that usually presents as syncope during late childhood or adolescence.
- Acquired long QT syndrome can be associated with hypokalemia, hypomagnesemia, severe malnutrition, and intracranial catastrophes such as subarachnoid hemorrhage
- Family history of sudden death
- Congenital deafness
- Unexplained syncope
- Previous arrhythmias
- Physical exam dictated primarily by surgical procedure and co-morbid medical conditions

INVESTIGATIONS

- Electrolytes (including K^+, Ca^{2+} and Mg^{2+})
- EKG (The strongest predictor of the risk of syncope or sudden death in patients with congenital prolonged QT syndrome is a QTc exceeding 500 milliseconds)

OPTIMIZATION

- Consider pre-operative β-blockade or prophylactic LEFT stellate ganglion block
- Correct electrolyte abnormalities (K^+, Mg^{2+}, Ca^{2+})
- Consider placing cardioversion/defibrillator pads on patient prior to draping
- Avoid bradycardia
- Avoid sympathetic stimulation
- Any drugs associated with QT prolongation should be discontinued

ANESTHETIC OPTIONS

- Regional, neuroaxial, and GA suitable

ANESTHETIC SETUP

- **Equipment**
    - Standard equipment
    - Defibrillator / Crash cart
- **Monitors**
    - Standard CAS monitors + 5 lead EKG
    - Adequate IV access (for resuscitation if required)
    - Invasive monitors only if required by surgery or other co-morbidities
- **Drugs**
    - Standard drugs including resuscitative drugs (phenylephrine and ephedrine)
    - Mg^{2+}
    - ACLS drugs
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MANAGEMENT OF ANESTHESIA

- **Premedication**
    - Anxiolysis to reduce SNS discharge
- **Induction**
    - Avoid Ketamine and Pancuronium given sympathomimetic activity

- Propofol has a reported ability to shorten QT interval → may be beneficial

- **Maintenance**
  - TIVA may be optimal choice
  - Halothane, Isoflurane, Sevoflurane all prolong QT interval but have been used safely in patients with prolonged QT
  - Consider having cardioversion pads on patient
  - Ensure adequate depth of anesthesia, so as to avoid SNS activation
  - Avoid drugs that prolong QT (e.g. droperidol, 5-HT3 antagonists)

COMPLICATIONS

- Perioperative ventricular tachycardia or fibrillation

PATHOPHYSIOLOGY

- The congenital form of the LQTS consists of defects in cardiac ion channels that are responsible for cardiac repolarization
- Defects that enhance Na^+ or Ca^{++} inward currents or inhibit outward K^+ currents during the plateau phase of the action potential lengthen the QT-interval
- Longer QT-interval predisposes to polymorphic ventricular arrhythmias in response to the triggers (exercise, emotional stress, sleep)
- Patients with acquired LQTS can develop marked QT prolongation in response to drugs that alter repolarization currents
- Drug-induced long QT and associated polymorphic ventricular tachycardia are frequently potentiated by development of hypokalemia and bradycardia

Table 3. Effects of various anaesthetic drugs on the QT interval.

	Prolongs QT interval	Does not prolong QT interval
Induction agents	Thiopental, ketamine	Propofol, etomidate, methohexital
Volatile anaesthetics	Isoflurane, sevoflurane	Halothane
Muscle relaxants	Succinylcholine, pancuronium	Vecuronium, atracurium
Opioids	Sufentanil	Alfentanil, fentanyl
Benzodiazepines		Midazolam
Drugs used to 'antagonize' residual neuromuscular blockade	Neostigmine, edrophonium, atropine, glycopyrrolate	
Sympathomimetics	Epinephrine, norepinephrine	Phenylephrine

REFERENCES

- Stoelting's Anesthesia and Co-Existing Disease, 5th Edition